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REPORT OF A CASE OF BRAIN AND SPINAL CORD SYPHILIS, SHOWING EXTENSIVE DISEASE OF THE BLOOD-VESSELS.

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WITH AN ACCOUNT OF THE AUTOPSY AND MICROSCOPIC EXAMINATION.

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A MAN, then 63 years of age, had in January or February, 1877, a macular syphilitic eruption upon the face, trunk, and limbs, and, at the same time, a violently painful iritis. From these disorders he recovered, but continued in rather an ailing condition until September 13th, when, after having dragged the right leg, and having been somewhat awkward in walking for about two weeks, he was seized with right hemiplegia. The paralysis was not complete, although very pronounced, for he was still able to walk a little; there was also decided thickness of speech, although at this time there was no absolute loss of power of speech. After this attack, he slowly improved, and was able to be up until about December 1st, when, after suffering with some dimness of vision and feeling of general *malaise* and staggering gait, he was seized with violent vomiting and a fresh access of paralysis, which was now almost complete, so that for a time he was unable to walk at all. Again he improved, but on May 13th, 1878, had another return of paralysis, which was complete, and there was entire loss of speech, except that he could say "yes" and "no." In the latter part of April or early in May, 1880, he had some vague increase of paralysis, and on May 23d had an attack, which was so sudden that he nearly fell, and afterwards complained of a strange sensation upon the left side. Upon investigation, it was found that there was almost complete loss of cutaneous sensibility in the left leg.

After each of his attacks he slowly improved, and was usually soon able to walk with assistance or even alone, until August 29th, 1881, when he fell upon the floor of his room and broke the neck of the left femur, after which, of course, he never walked again, but sat in a chair or lay in bed. Within two years and a half after the original onset of his illness, he had twitching of the right leg and trembling, which was often so violent as to shake the bed, and stiffening of the right leg came on as he was getting about after the attack of paralysis in May, 1880. May 3d, 1885, he was slightly out of his head, and had a constant tendency to fall over to the left side in his chair. June 7th, 1885, it was discovered that he had gangrene of the left heel, which soon extended to the toe, and then increased in extent until it involved about half of the foot. In August, a line of demarcation formed, and the gangrene never extended after that time.[1] There was general cutaneous hyperæsthesia for about two months before his death, which took place October 10th, 1885.

Post-mortem examination was made about thirty hours after death. The spinal cord was removed. Upon opening its membranes, nothing unusual was observed until the upper lumbar region was reached, when a mass was found with the appearance, upon section, of a cyst containing blood; this was attached to the dura mater upon the left side. The cord, upon section, at its upper portion seemed softer than usual, and the cut surface presented a pitted appearance. The degree of softening increased progressively from above downwards until the lower dorsal region was reached, at which position, upon section, it was seen to be almost semifluid. The lumbar cord in its upper portion was much broken down; below again it seemed much firmer. The softening was below the mass mentioned, and attained its maximum degree about three-quarters of an inch below the mass.

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[1] The gangrene was of the dry variety, and there never was the slightest odor connected with it.

On removing the skull cap, the dura mater was found to be tightly adherent to it throughout, so that it was removed with difficulty. On examining the dura mater, some spots seemed thicker than others, and there were two dark-reddish œdematous-looking spots upon the left side. Upon the surface of the brain corresponding to the described areas there were serous effusions in the arachnoid. The general surface of the brain presented no unusual appearance except that the convolutions looked flattened and narrowed; its surface seemed rather firmer than usual. The pia mater could be stripped from the brain at any portion with perfect ease, coming out without difficulty even from the deeper sulci, and showing the size and outlines of the convolutions. The inferior surface of the brain, when examined from before backwards, presented nothing of note until the crura cerebri were reached, when it was seen that the left crus was universally darker in color than the right, its inner edge being for about one-fourth of an inch dark slate-colored and ill-defined. Upon section, above the corpus callosum on the left side the white matter appeared to be normal. When the left lateral ventricle was opened, there were seen upon the surface of the corpus striatum two areas of softened matter which were dark gray in color and almost translucent and very soft. The optic thalamus also showed two spots of softening. Upon section of the corpus striatum, its middle portion was found to be converted into a cavity containing serous fluid, although the anterior and posterior portions looked more natural. The inner portion of the optic thalamus was soft and reddish. The right corpus striatum and optic thalamus seemed to be natural.

When the thorax and abdomen were opened, it was seen that the amount of adipose tissue was unusually great, both in the walls of the cavities and in the omentum, and the cavities seemed to be too large for the organs, which were all shrunken and small. The lungs crepitated throughout, and there were no adhesions upon the right side. The left lung was somewhat adherent at its posterior middle surface. The heart was small, and its tissue very dark colored. The aortic opening contained calcareous deposits which were most marked upon the intercoronary flap of the valve. The aorta was about twice the natural size, being atheromatous and stiff, but not pouchy. The mitral curtains were somewhat thickened, but otherwise healthy. The liver was very small, and of a dark nutmeg color; its surface rough, and the capsule thickened. Upon section, the tissue of the liver for a quarter of an inch beneath the capsule was seen to be lighter colored than the deeper portion, which was very dark. The gall-bladder was adherent and thickened, and contained a gall-stone three-quarters of an inch in diameter. The kidneys were small and had a markedly urinous odor, and were dark nutmeg-colored. The capsules were thickened and removed with difficulty, and the veins upon the surface were well marked.

The left femur was found to have been fractured at the neck, partly within and partly without the joint, and there was much shortening of the bone, and the seat of fracture was surrounded with a large amount of callus.

REPORT OF MICROSCOPICAL EXAMINATION.

Heart.—The muscular fibres were apparently smaller and narrower than normal, although this was not determined by any comparative measurements. Quite numerous young connective-tissue cells appeared between the fibres. Small deposits of yellowish-brown pigment were present in the muscle fibres, chiefly situated at either pole of their nuclei.

Kidney.—There was an increased amount of intertubular connective tissue, and often rich collections of dark-stained nuclei. The arterioles had thickened walls, especially the muscular coats, while the increased fibrous coats gradually united with connective tissue around, which was often well supplied with nuclei. The cells of the uriniferous tubules were granular and indistinct. The process was the early stage of interstitial nephritis.

Liver.—The lesions of a well-marked fatty infiltration were present. Slight increase of fibrous tissue, and in many acini indistinct and really granular cells were also present.

Nervous System.—Parts taken for examination: third left frontal convolution, left optic thalamus and corpus striatum, left crus cerebri, right optic thalamus, pons, medulla, spinal cord, and small tumor attached to meninges opposite the upper lumbar cord.

The specimens were hardened for section by placing them for eight weeks in a mixture of three parts of Müller's fluid to one of methylic spirit, with frequent changing then in dilute alcohol for one week, and finally for forty-eight hours in absolute alcohol.

These portions were permeated with paraffin, sections cut and pasted on the slides, the paraffin dissolved, and the sections stained in situ, thus reducing any distortion of the relations of the parts to a minimum. The stains were carmine, picro-carmine, and the double stains carmine and sulpho-indigotate of potash. The finer sections from which this study has been made were cut by Dr. Wm. M. Gray, microscopist of the Army Medical Museum.

Convolution.—There was a general enlargement of the capillaries, which were distended with blood, and surrounded by dilated lymph spaces. The larger vessels were similarly engorged with blood, and in places rupture of the wall had taken place and allowed an escape of blood-corpuscles into the surrounding tissue. The arterioles presented distinctly thickened walls—the thickening was of the coats outside the intima, and each vessel was surrounded by an open space caused by absorption and atrophy of the surrounding tissue. The edges of these spaces were frayed and usually more deeply tinted than the adjoining structures, and in the immediate neighborhood round and oval bodies, probably amyloid, were often noticeable. The ganglion cells were cloudy, often showed beginning pigmentation and poor staining, and in places were positively shrunken.

Left Optic Thalamus and Corpus Striatum.—There was an extensive degeneration of the normal elements. Such ganglion cells as remained were shrunken and cloudy, while in the major portion of the diseased area there was an entire absence of nervous elements, which had been displaced by numerous bands and bundles of sclerotic tissue that crossed and recrossed one another, leaving intervening spaces until the whole resembled a coarse lace-work texture. The blood-vessels were distended with blood, and showed thickened walls. In the less diseased parts there were collections of small round cells and a very general increase in the neuroglia.

Left Crus Cerebri.—The cells of the substantia nigrica showed the usual dense pigmentation, perhaps more so than normal. Their processes were shortened or absent. Anterior and posterior to this region distended vessels—sometimes with thickened walls, and here and there in relation to extravasations of corpuscles—were noted.

Right Corpus Striatum, Medulla, and Pons.—Appearances similar to what have been described were present in less marked degree.

Spinal Cord (cervical and dorsal).—The central canal did not exist as a channel, but was plugged up by its proliferated epithelial elements. In its immediate neighborhood, in the gray matter, extensive thickening of the walls of the smaller arterioles and capillaries existed, producing the appearances of *hyaline degeneration.* This pathological change was present in all degrees from the slightest thickening up to aggregated masses which surrounded the vessel tube. The ganglion cells were ill-defined, shrunken, unequally stained, and often markedly pigmented. Their processes were almost universally shortened and frequently absent. In addition to spaces around the enlarged vessels, radiating fissures occurred, especially in the posterior gray horns, having no relation to vessel walls. No positive change in the nerve tubules of the white matter was present.

The membranes of the cord showed in places collections of inflammatory cells and vessels with thickened walls.

Tumor.—This was composed of an external envelope of well-felted, wavy connective tissue, through which a few young cells were scattered. Internally were the remains of a tissue composed partly of small round cells, like granulation tissue, and partly of blood-corpuscles and yellow pigment granules. The growth was probably a gumma.

Cord Opposite the Tumor.—The cord here showed extensive destructive changes. Nerve-cells and tubes were destroyed, wasted, and pressed apart. Numerous round and oval bodies were scattered through the degenerated tissue, which may have been amyloid, or, as Kesteven has suggested, ends of broken nerve-fibres. The hyaline change in the vessel walls was here seen in its highest perfection, so much so as, in some instances, to have entirely occluded the vessel lumen.

Résumé and Remarks.—From this study it is evident that the lesions may be arranged as follows:

1. Coarse, destructive disease of all tissues of the left corpus striatum and optic thalamus, and of the lumbar cord where pressed upon by the tumor.

2. A very general lesion of the blood-vessels, which manifested itself in the brain by dilatation of the capillaries and thickening of the walls of the larger vessels, and in the spinal cord, especially low down, by a typical *hyaline* degeneration of the capillaries and smaller arterioles.

3. Scattered areas of degeneration, which were either in relation to enlarged blood-vessels, and contained blood-corpuscles which had escaped through rupture, or had no such relations, and have been described as radiating fis-

sures. The exact significance of these fissures is doubtful, and they may have been caused by the process of handling or hardening the specimen. Although Kesteven [1] has described " radiating fissures which result from loss of the connective which, in the normal condition, binds together the nerve fibres into bundles," it seems likely, in the present instance, they had no pathological significance. Not so with the areas relating to blood-vessels, which, at least in many spots, had undoubted pathological importance.

4. A quite universal change of the ganglion cells, which were cloudy, unequally stained, pigmented, and had their processes either shortened or lost. This appearance is not always as important as it at first thought seems, especially when it is remembered that the recent experiments of F. Kreyssig [2] show that poor staining and even vacuolation of cells may occur from putting tissues hardened in chromates directly into strong alcohol. This method had been avoided in the present case, and hence it seems fair to suppose the lesion had pathological significance.

Remarks.—The features of this case that seemed to make it worth reporting were that the man was under observation from the time of the onset of the first symptoms of secondary syphilis to his death, eight and a half years afterwards, and that there was an opportunity to make the most extended investigation of the post-mortem conditions. The history of the patient is sufficiently exact and minute with regard to the dates and mode of onset of the various attacks, but many of the finer methods of diagnosis were not made use of, as, for instance, any study of the electrical reactions and the more delicate methods of examining the conditions of the organs of special sense.

It is in a high degree interesting to compare the symptoms as they were observed during life in this case with the post-mortem appearances. There were three distinct attacks of hemiplegia, one accompanied with aphasia, and after each one great amelioration of the symptoms took place; then came on secondary muscular twitching and contracture, and finally gradual loss of intellectual power. It seems hardly likely that the cause of these successive attacks could have been the condition of the left corpus striatum, for the disorganization of that portion of the brain substance was so complete that most probably this was of comparatively recent origin, and the attacks of hemiplegia which occurred seven years before death were due to some other cause. The only other possible cause discovered was the minute extravasations of blood which, in certain areas, were found in the brain substance around the arterioles. The old disintegrated sac, which was found low down in the spinal canal, was probably all that remained of a gumma which had softened down under the administration of the large doses of iodide of potassium which this patient took for a long time. This tumor and the secondary softening of the cord which existed below it were the only coarse syphilitic lesions which were discovered. There must, of course, have been paraplegia as a consequence of this, but it was unobserved, owing to the multitude of other causes that existed to make the limbs motionless. It cannot be known at what period of the patient's illness this tumor came on, but this is a matter of no moment, for there was no true paraplegia either at the time when paralysis first came on or for a long time afterwards, as the patient continued to walk a little until he had the fall which has been mentioned, and which resulted in a fracture of the neck of the femur.

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[1] "Observation of some points in the Path. Histol. of Spinal Cord." Trans. Internat. Med. Congress, 1881, Vol. I.

[2] Virch. Archiv, Bd. 12.

The fact is a most striking one, that, although at first sight Broca's convolution seemed to present no evidence of disease, outside of its partaking of the shrunken and hardened condition which was everywhere evident upon examination of the brain surface, yet the microscope revealed the existence of a condition of the arterial supply which, upon careful consideration, was exactly what might have been expected when the state of the patient's power of speech and mind were recalled. After the violence of the hemiplegic attack with aphasia, which occurred more than six years before his death, had passed off, the power of speech seemed nearly normal, but from that time it gradually declined until, when he died, he had certainly not more than one-fourth the natural power of communicating his ideas by speech. His word memory was very poor, and he had great difficulty both in getting a mental grasp upon the word he was in need of and then in enunciating it, and his sentences were always short and ill-constructed, like those of a young child. At the same time that the aphasia gradually increased, it may almost be said that his intellectual powers "sclerosed"—he seemed to have less thought and less feeling, and became all the time less and less like a man, and more and more like an animal. Naturally a man of few words, he became positively morose in his silence, sitting for hours in his chair, glowering at nothing if undisturbed, and snarling a monosyllabic answer if asked a question. Still he had his pleasures, and one of his greatest was to listen to any conversation that might be going on within his hearing. Although nothing ever induced him to join in it, either voluntarily or if asked, still he would often show that his power of memory was fairly good by letting it be known that he recollected most of what had been said. This condition of almost complete aphasia, as it finally became, would seem to offer very strong evidence, if any further evidence is needed at this day, of the correctness of locating the speech centre in Broca's convolution, for the way in which the power of speech gradually declined through years, and the way in which the blood supply must have been gradually choked off by the slowly increasing vascular disease were most strikingly in accord.

No thoroughly satisfactory explanation of the cause of the successive attacks of paralysis, which are so common in cases of brain syphilis, seems as yet to have been suggested by pathologists, and yet it is one of the most marked clinical features of the disease. Syphilitic paralysis seldom comes on with the same suddenness as that caused by hemorrhage or the washing of an embolus into one of the cerebral arteries, nor is it usually so complete or of such long duration. There is commonly some warning of the onset of an attack of syphilitic paralysis—for some hours or even for a day before, the patient feels strangely, or has headache, or there is partial paralysis of the part subsequently to become completely so, and this fact constitutes usually one of the most valuable and reliable signs to enable the physician to make a differential diagnosis between syphilitic and other forms of paralysis. It may possibly be that the explanation of the recurring attacks of hemiplegia in this case is to be found in the small extravasations which existed near the blood-vessels in some areas of the motor tract.

The general hyperæsthesia, which was a marked feature in the last few weeks, was fully accounted for by the meningitis which the post-mortem examination revealed. The existence of an unusually large bed-sore over the sacrum coincidently with the meningitis and disease of the cord, which has been described, would seem to go to show that it was due to trophic nerve influence, as it has been described by Charcot, although the likelihood of trophic influences being capable of producing such an effect has been doubted by Green.[1] It is a thing not very uncommon for old people, or those who have aged prematurely, to have general hyperæsthesia and bed-sores as the last chapter of some wasting condition which finally ends their lives, and this goes toward showing the hyperæsthesia and the meningitis, which certainly caused it in this case, to have had some more than casual relation with the occurrence of bed-sore. This mode of death of old people does not seem to be commonly known among clinical observers.

The striking and unusual features of the case may be briefly summed up as follows:

1. The fact that Broca's convolution, which, upon macroscopical examination, presented no evidence of disease, was found to have a degree of change in and around the blood-vessels which fully accounted for the aphasia.

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[1] "Pathology and Morbid Anatomy," by T. Henry Green, American edition, page 26.

2. The exact correspondence there was between the clinical history of the case, in regard to the constantly increasing aphasia and the slow decrease of the intellectual powers, and the condition of the blood-vessel system discovered upon microscopic examination—for the supply of blood must have been slowly cut off from year to year, thus gradually producing the results observed.

3. The possibility that the successive attacks of hemiplegia which occurred in the case, and which were always preceded by premonitory symptoms, as is usual in cases of brain syphilis, may have been due to the extravasations which were found around the blood-vessels at various areas in the motor tract.